

Taste and olfactory status in a gourmand with a right amygdala lesion

M. Gallo¹, F. Gámiz¹, M. Perez-García^{1,2}, R. G. del Moral³, and E. T. Rolls⁴

¹Department of Psychobiology, Instituto de Neurociencias F. Olóriz, Centro de Investigaciones Biomédicas, CIBM, Universidad de Granada, Granada, Spain

²Centro de Investigaciones Biomédicas en Red de Salud Mental, CIBERSAM, Universidad de Granada, Granada, Spain

³Hospital Universitario San Cecilio, Universidad de Granada, Granada, Spain

⁴Oxford Centre for Computational Neuroscience, Oxford, UK

In a patient with a lesion of the right amygdala and temporal pole who had the characteristics of the gourmand syndrome, sensory and hedonic testing was performed to examine the processing of taste, olfactory, and some emotional stimuli. The gourmand syndrome describes a preoccupation with food and a preference for fine eating and is associated with right anterior lesions. It was found that the taste thresholds for sweet, salt, bitter, and sour were normal; that the patient did not dislike the taste of salt (NaCl) at low and moderate concentrations as much as age-matched controls; that this also occurred for monosodium glutamate (MSG); that there were some olfactory differences from normal controls; and that there was a marked reduction in the ability to detect face expressions of disgust.

Keywords: Gourmand syndrome; Amygdala; Anterior temporal lobe; Taste; Olfaction; Disgust.

In this article, we describe sensory and hedonic taste and olfactory tests, and neuropsychological investigation, in a person who became a gourmand associated with a meningioma in the sphenoid bone which produced damage in the right anterior temporal lobe of the brain, particularly in the amygdala. The gourmand syndrome describes a preoccupation with food and a preference for fine eating and is associated with right anterior lesions of the brain involving cortical areas, basal ganglia, or limbic structures (Regard & Landis, 1997). Part

of the aim was to investigate whether any changes in the taste and olfactory systems were associated with the gourmand status of the person. Previous publications have not involved olfactory and taste testing of patients with the gourmand syndrome and have not described damage in the amygdala and temporal pole in particular (Cockrell, 1998; Kurian et al., 2008; Myslobodsky, 2003; Regard & Landis, 1997). Some of the previous cases have had damage more posteriorly in the temporal lobe, e.g., in the posterior ventral insula.

Address correspondence to Edmund T. Rolls, Oxford Centre for Computational Neuroscience, Oxford, UK and Department of Computer Science, University of Warwick, Coventry CV4 7AL, UK. (E-mail: Edmund.Rolls@oxcns.org).

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CASE REPORT

Medical history

RG is a right-handed male aged 60 with no history of eating disorders in his family. At the age of 42, his interest in gastronomy started, and his interest in running (he was a marathon runner) declined. He bought all the world gastronomy guides available and planned visits to the best-known restaurants, spending a lot of money. He even arranged long-distance travels abroad by car with the only intention of stopping to have dinner in famous restaurants. This is a typical gourmet behavior that has been termed “gastronomadism” (traveling hundreds of kilometers just to eat refined food). RG started to write summaries of his gastronomic experiences. RG’s history shows no evidence during childhood, adolescence, and previous adulthood of having been exposed to refined cooking or gastronomic habits. Instead, he reports to have considered the best food before this time to be that cooked by his mother, whereas gourmards typically reject everyday food and look for new gastronomic experiences continuously. Further, RG’s wife confirms a sudden change in his interest in gastronomy from the age of 42. At the age of 46, RG had an olfactory (not taste) hallucination for the first time: he experienced the smell of the cooking by his mother. This lasted for 2 minutes. It was pleasant at first, and then became unpleasant, a “chemical” smell. This recurred one month later, when he was in bed. When it recurred a third time, RG consulted a neurologist, and an MRI scan showed a fibroblastic meningioma in the minor wing of the sphenoid bone pressing on the right temporal lobe. The meningioma was removed in a 14-hour operation in 1997 at age 46. There was a brief ischemic episode during the surgery, while under the anesthesia. There were no more olfactory hallucinations after the surgery.

At the age of 47, RG became, in addition to his professional post, a restaurant gastronomic critic, and by the age of 54 was widely recognized as a gastronomic critic and wrote for prestigious gastronomic guides, newspapers, etc. He became overweight, gaining 50 kg. At that time, the gourmand behavior of the patient fully matched that described by Regard and Landis (1997) in their seminal paper, i.e., an abnormal and obsessive preoccupation for food; passion and preference for fine eating; and craving and expectation of good food. At the age of 58 (3 March 2010), with RG still a gourmand, a (routine) follow-up MRI scan was performed. The

main small brain region with damage evident was the right amygdala, in its dorsolateral part, and the right temporal pole (Figure 1a–c). No damage was evident in the orbitofrontal cortex, insular taste cortex, or hippocampus. In addition to this brain damage, presumably caused by the space-occupying meningioma detected and then removed in 1997, some tumor tissues were detected and were associated with the cavernous sinus, and radiosurgery was used to treat the tissue associated with the cavernous sinus (on 14 July 2010). There was no feeling of sickness or being unwell. Three days later, RG had some emesis due to brain swelling. An antiemetic was prescribed. The growth associated with the cavernous sinus had been pressing on the medial temporal lobe, medial to the amygdala, and had a diameter of 3.5 mm prior to the radiosurgery (see Figure 1d). After the radiosurgery, the diameter was approximately 2.5 mm, and it was less clearly pressing on the medial temporal lobe.

One month after surgery, RG lost his interest, his motivation, in gastronomy. As an example, he reports that at the time of receiving radiotherapy at a different town he organized a longer trip than needed in order to visit several high-level restaurants, both before and immediately after the intervention. However, one month later, he found the fine-quality food of the restaurants that he visited as gastronomic critic boring and was not interested in the food. He also experienced a personality change with blunted emotions. His wife recognized a change, explaining that he seemed to be a different person than before the radiotherapy. He remained critical in his evaluation of food. He did not detect a change in his interest in art, music, etc. He reported no problems with his memory and continues his professional activity with great success. MRI scans performed one year after the radiotherapy intervention showed that the damage in the dorsolateral part of the right amygdala and in the temporal pole was still evident and that the growth associated with the cavernous sinus remained small.

At age 60 (16 months after radiotherapy), the olfactory, taste, and neuropsychological testing described here was performed. At that time he had lost the overweight gained before the radiation therapy and was on a diet.

Pathology

The pathology in the brain was to the right amygdala, with some damage also to the right temporal pole cortex just anterior to the amygdala.

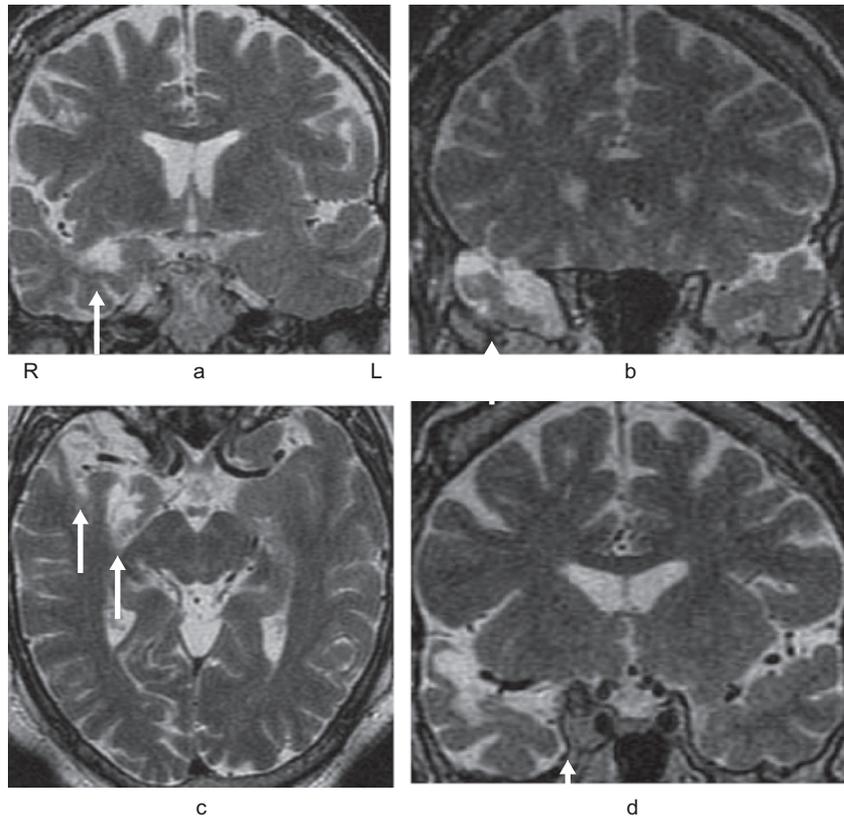


Figure 1. T2 MRI scans of brain lesion (14 July 2010). a. Coronal plane showing amygdala. b. Coronal plane showing temporal pole. c. Axial plane showing temporal pole and amygdala. d. Coronal plane showing tissue associated with the cavernous sinus. The arrows indicate the regions to which reference is made.

The hippocampus and insula were remarkably intact. The lesion is illustrated in Figure 1. The scans shown were obtained on 14 July 2010 immediately before the radiotherapy, and similar scans were obtained on 3 March 2010 and on 5 July 2011. The evidence thus suggests that the lesion has been present since the meningioma was detected in 1997. There is an indication from the scans in 1997 of the growth associated with the cavernous sinus, which is shown in the 2010 scan in Figure 1d. Soon after this region, shown in Figure 1d, received radiotherapy in July 2010, the gourmand status declined.

Self-report

RG describes his gourmand skill as being intellectual. He has great motivation to describe food and believes that this rather than any hypersensitivity is important in his passion for food. RG very much likes endives and tonic water. RG thinks that he has

a high threshold for quinine. RG uses saccharine, so he thinks he may need a lot of sucrose during the sensory testing to like it. In his taste subjective ratings described below, he found low concentrations of sucrose not very pleasant and said that this was because they are weak to him.

Sensory testing

Taste thresholds

Taste thresholds to sweet (sucrose), salt (NaCl), bitter (quinine HCl), and sour (HCl) were measured using the methods described by Donaldson and colleagues (Heath, Melichar, Nutt, & Donaldson, 2006). Tastants were provided for each trial in 1 ml amounts with the instruction to move this round the mouth and to indicate whether a taste was detected. The subject was not told what taste was being presented on each trial. Between each stimulus application, a 30 s interstimulus interval was adhered to, during which time the subject would

rinse the mouth with 5 ml of deionized water. Each concentration of stimulus was presented to the subject 3–5 times in total (with testing spread over several days in 1 hour sessions with a rest at 30 min), and the percentage of positive responses at each concentration could therefore be determined. The subject was first presented with a solution that should be above threshold. Thereafter, stimuli were presented in a pseudorandom order in concentrations representing 1/4 log steps between the lowest (undetectable, 0% detection) and the highest (always detectable, 100% detection) concentrations. This protocol was adopted to minimize both adaptation to the stimulus and guessing by the subject. The range of concentrations varied for different modalities as follows: sweet (300 mM to 0.3 mM sucrose), bitter (300–0.3 μ M quinine hydrochloride), salt (100–0.3 mM NaCl), sour (100–0.55 mM HCl), and umami (100–0.3 mM MSG). Solutions were prepared with deionized water shortly before testing and were presented at room temperature. Taste psychometric functions based on the percentage of positive taste recognition against log solute concentration were generated (see Figure 2). From these curves, taste recognition thresholds (the concentration at which the subject would recognize the taste 50% of the time) were calculated. Standard fitted sigmoidal stimulus–response curves of the percentage of correct taste identification versus \log_{10} tastant concentration (molar) were used to measure sensory thresholds.

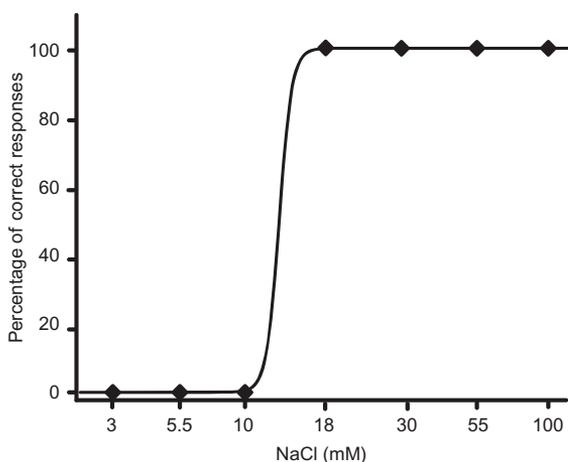


Figure 2. RG's taste threshold data for NaCl. The mean percentage correct (\pm SE) as a function of the concentration of NaCl (on a log scale) is shown. A sigmoid was fitted to the data. The threshold was taken at the concentration at which the detection of the taste was 50% correct.

Affective and intensity ratings of taste stimuli

Most foods have concentrations of tastants that are well above threshold, and pleasantness and intensity ratings at suprathreshold concentrations are useful in investigating factors that influence food liking and intake. For example, the pleasantness or reward value of tastes and odors, but not their intensity, is reduced by consuming a food to satiety (Rolls & Rolls, 1997; Rolls, Rolls, & Rowe, 1983). Further, the pleasantness and intensity of tastes are represented separately in the brain (Rolls, 2014), in that activations in the insular primary taste cortex are correlated with the subjective intensity of taste (Grabenhorst & Rolls, 2008) and reflect the concentration (Grabenhorst, Rolls, & Bilderbeck, 2008), whereas activations in the orbitofrontal cortex are correlated with the subjective pleasantness of a taste (Grabenhorst & Rolls, 2008) and decrease when flavor pleasantness is decreased by feeding to satiety (Kringelbach, O'Doherty, Rolls, & Andrews, 2003). For these reasons, we measured the subjective intensity and pleasantness of a range of concentrations of selected taste stimuli to investigate the processing of taste at concentrations found in foods.

The protocol and stimuli followed the procedure of Rolls et al. (1983), with the data obtained in that investigation used for comparison. The pleasantness and intensity of the taste of solutions of sucrose (0.1, 0.2, 0.4, 0.8, and 1.6 M) and of salt (0.1, 0.2, 0.4, 0.8, and 1.6 M NaCl) were rated using 100 mm visual analog rating scales. For intensity, the ends were marked Very Weak and Very Intense, and measurements were in mm from the end marked Very Weak. For pleasantness, the ends were marked Very Unpleasant and Very Pleasant, and measurements were in mm from the end marked Very Unpleasant. A mark was made at the point on each scale to represent the rating of the 1 ml of solution provided on each trial. The solutions were presented in random order, separated by a rinse with 5 ml of water, until three ratings for each solution had been made. The use of these rating scales has been validated in previous studies and is useful especially when within-subject comparisons are made, e.g., for a given subject how the pleasantness and intensity are influenced by feeding to satiety (Rolls et al., 1983). In the present context, a within-subject comparison can be made for sucrose versus salt, for comparison with previous data (Rolls et al., 1983).

Smell identification: UPSIT, long version

To assess RG's olfactory status, the University of Pennsylvania Smell Identification Test (UPSIT, Sensonics Inc., Haddonfield, NJ, USA (Doty, 2008; Doty, Shaman, & Dann, 1984)) was performed.

Olfactory psychophysics

As described above, ratings of the pleasantness and intensity of odors at above-threshold concentrations may be relevant to normal olfactory function. It is important to assess pleasantness as well as intensity, for the pleasantness but much less the intensity of an odor is reduced by feeding it to satiety (Rolls & Rolls, 1997), and the pleasantness or reward value of odor is relevant to appetite and the control of food intake (Rolls, 2012, 2014). Consistently, there are separate representations in the brain, with activity in the orbitofrontal cortex reflecting the reward value and pleasantness of odors (Critchley & Rolls, 1996; Grabenhorst, Rolls, Margot, da Silva, & Velazco, 2007) and activity in the pyriform (primary olfactory) cortex reflecting the intensity of odors (Grabenhorst et al., 2007). To address whether there might be a difference in pleasantness and/or intensity between RG and controls, we devised a new test in which we also measured RG's pleasantness and intensity ratings for the items in the UPSIT (long) and compared these with the ratings (\pm SEM) of six age-matched controls from the same department. The same visual analog rating scales were used as for the taste subjective ratings.

In addition, RG's olfactory threshold was also measured with phenylethylamine (Smell Threshold Test, Sensonics Inc. (Doty, 2008; Doty, Gregor, & Settle, 1986; Doty, Kisat, & Tourbier, 2008; Doty & Laing, 2003; Doty, McKeown, Lee, & Shaman, 1995)). In the protocol, a two-alternative forced-choice single staircase procedure was used to establish detection threshold values for the rose-like smelling odorant phenyl ethyl alcohol (PEA). The staircase began at the -6.0 log concentration step of a half-log step (vol/vol) dilution series extending from -10.0 log to -2.0 log concentration. The stimulus concentration was increased in full log steps until correct detection occurred on five sets of consecutive trials at a given concentration. If an incorrect response was given on any trial, the staircase was moved upward one full log step. When a correct response was made on all five trials, the staircase was reversed and subsequently moved up or down in 0.5 log increments

or decrements, depending upon the subject's performance on two pairs of trials at each concentration step. The geometric mean of four staircase reversal points following the third staircase reversal was used as the threshold measure. The test-retest reliability of this instrument is >0.80 (Doty et al., 1995).

Emotion perception

Because it was evident that RG had an amygdala lesion, it was of interest to test whether there might be any changes in some of the functions of the amygdala in emotion, and this was of special interest as RG has unilateral damage, not the bilateral damage that is the subject of some previous investigations. The ability to identify face expressions was of particular interest as deficits in the ability to correctly identify face expressions of fear have been reported previously (Adolphs et al., 2005; Calder et al., 1996; Phelps & LeDoux, 2005). The ability to recognize face expressions was measured with the Facial Expression of Emotion: Stimuli and Tests (FEEST) (Young, Perrett, Calder, Sprengelmeyer, & Ekman, 2002) assessment for emotion perception. FEEST comprises two tests: the 60 Faces Test and Emotion Hexagon Test. In the former test, a total of 60 photos of faces are presented using the CD software in a random order for five seconds each, with 10 photos for each of the six basic emotions from the Ekman and Friesen (1976) series (happiness, surprise, fear, sadness, disgust, and anger). Participants had a choice of six emotion labels: "happiness," "sadness," "anger," "disgust," "fear," and "surprise." Ten trials for each emotion were presented in random order, and the participants received no feedback on task performance. The Emotion Hexagon Test uses computer image manipulation techniques to test facial expression recognition with stimuli of graded difficulty. The computer software on the CD-ROM presents the stimuli in random order for 5 seconds each across one practice and 5 test blocks of 30 trials each, and records responses made from mouse clicks to on-screen buttons. Using Ekman and Friesen's (1976) norms, FEEST plotted a confusion matrix for the different emotions and then ordered them in a series based on their maximum confusabilities—placing each emotion adjacent to the one it was most likely to be confused with. The result ran happiness—surprise—fear—sadness—disgust—anger, with mean percentage confusabilities for each pair of expressions in this

sequence being happiness and surprise 0.8%, surprise and fear 5.8%, fear and sadness 2.4%, sadness and disgust 2.7%, and disgust and anger 6.4%. The ends of the sequence (anger and happiness) were then joined to create a hexagonal representation. Norms for the test were applied (Young et al., 2002).

RESULTS

Taste thresholds

Data of the type illustrated in Figure 2 showed that RG's taste thresholds were as follows: sucrose 3 mM; NaCl 15 mM; quinine 5.5 μ M; HCl 1.8 mM; MSG 1 mM. He is a taster of the bitter substance PROP (6-n-propylthiouracil). These thresholds compare to the following in a large group of subjects: sucrose 23 mM; NaCl 19 mM; quinine 30 μ M; HCl 10 mM (Heath et al., 2006).

RG's taste thresholds thus are in several cases lower than in normal controls, but this is probably due to the fact that in the investigation by Heath et al. (2006) approximately 0.2 ml of tastant was placed on the tongue with a cotton bud, whereas in the present investigation 1 ml was provided for whole mouth stimulation, and the latter is known to result in measured thresholds that can be ten times lower (Frank, Hettinger, Barry, Gent, & Doty, 2003). Nevertheless, RG's taste thresholds indicate excellent sensitivity to these four tastes, and this is especially the case in that taste thresholds may rise with age (Frank et al., 2003). RG's very good taste thresholds may relate to his expertise as a gourmand.

Taste psychophysics

RG's ratings of the pleasantness and intensity of sucrose and salt as a function of concentration are shown in Figure 3a. It was of interest that RG did

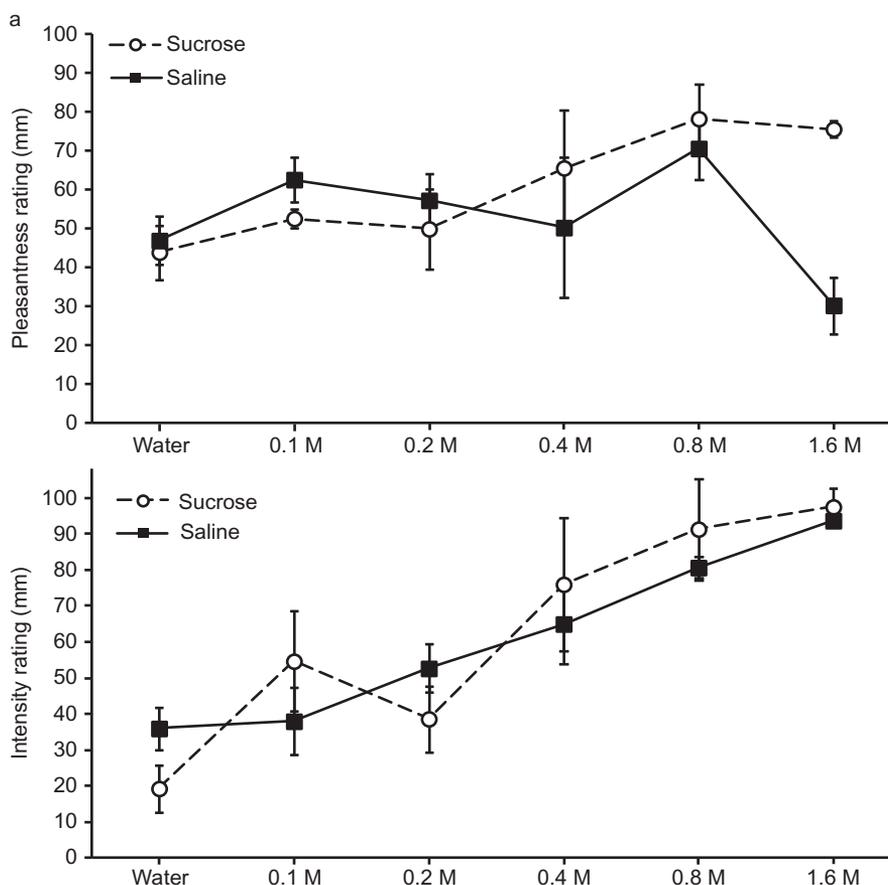


Figure 3 (above and next page). (a) RG's ratings of the pleasantness and intensity of sucrose and salt as a function of concentration. (b) Control subjects' ratings of the pleasantness and intensity of sucrose and salt as a function of concentration. Copyright © 1983, Elsevier. Reproduced with permission. Sensory-specific and motivation-specific satiety for the sight and taste of food and water in man. Rolls, E. T., Rolls, B. J., & Rowe, E. A., *Physiology and Behavior*, Vol 30, 185–192.

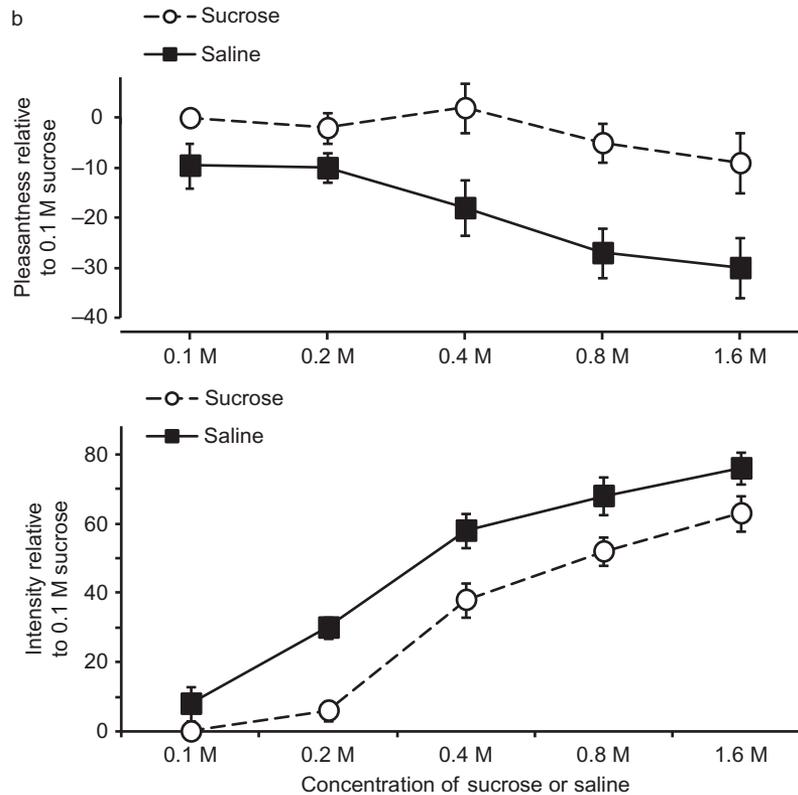


Figure 3. (Continued).

not find that sucrose was always, for a given concentration, more pleasant than salt. He even preferred the low concentrations of salt to sucrose. That is quite unusual, by a comparison with the ratings in a previous study (Figure 3b) (Rolls et al., 1983).

Although RG's intensity ratings of both sucrose and salt increased reasonably with concentration, it was interesting that his ratings of the intensity of the salt were in general not greater than those for sucrose at equimolar concentrations in contrast to those of previous control participants (Rolls et al., 1983). His relative preference for the lower concentrations of salt compared to sucrose was thus associated with a relatively low rating in the intensity of salt.

In view of these unusual ratings by RG, we performed further tests of his pleasantness and intensity ratings of further tastants and compared them with the ratings of a group of four age-matched controls who were professional colleagues from the same department. The results are shown in Figure 4. The ratings of the pleasantness shown in Figure 4 do indeed confirm that RG is unusual in liking the taste of NaCl and of monosodium

glutamate (MSG) more than the age-matched controls. On the other hand, his taste intensity ratings were not markedly different from those of the age-matched controls (Figure 4).

In summary, the taste psychophysics showed that RG had an unusual liking for NaCl at low and moderate concentrations, or, to put it another way, did not dislike the taste of salt as much as age-matched controls.

Smell identification: UPSIT, long version

RG's result was 33 (out of 40) on the UPSIT (Sensonics Inc. (Doty, 2008; Doty et al., 1984)). Using the validated scale of the administration manual (Doty, 2008) indicated a percentile score on the edge of a mild microsmia. This score compares to 36.16 with SD 1.32 for age-matched controls. This does not seem to be a marked reduction in smell identification.

Olfactory psychophysics

We also measured RG's pleasantness and intensity ratings for the items in the UPSIT (long) and

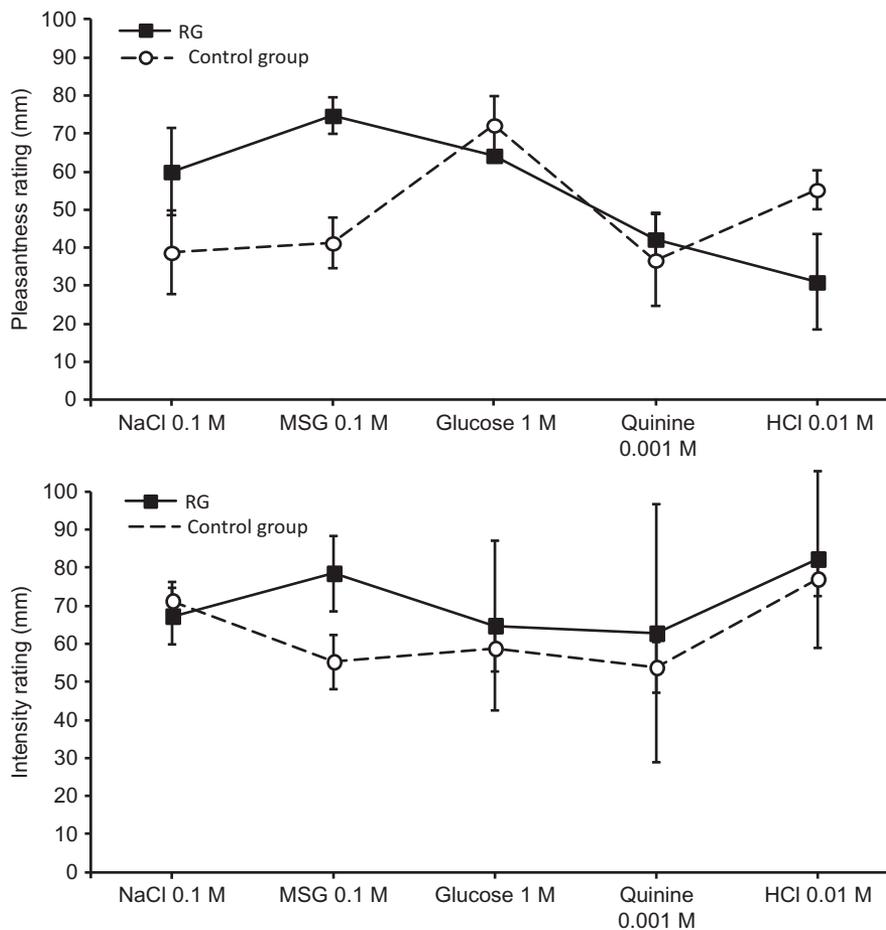


Figure 4. RG's ratings of the pleasantness and intensity of different tastes at standard concentrations are shown, together with data for comparison from four age-matched control subjects.

compared these with the ratings (\pm SEM) of age-matched controls in Table 1 ($n = 6$, average age = 58). The ratings were measured using a 100 mm visual analog rating scale. For pleasantness, the ends of the scale were marked "Very Unpleasant" and "Very Pleasant," and the measurement was in mm from the end labeled "Very Unpleasant." For intensity, the ends of the scale were marked "Very Weak" and "Very Intense," and the measurement was in mm from the end labeled "Very Weak." We note that a rating of more than two standard errors from the mean corresponds to $p < .05$. Items in which RG's ratings were more than two standard errors from the controls are marked with * in Table 1.

Odors that RG found less pleasant than the control group included menthol, banana, lilac, turpentine, pine, and artificial grapes. Odors that he found less intense than the control group included bubble gum, menthol, and clove. Odors that he found more

intense than the control group included orange, chocolate, nut, turpentine, and rose. There seemed to be no clear pattern to any possible differences in olfaction from age-matched controls.

RG's olfactory threshold was also measured with phenylethylamine (Smell Threshold Test, Sensonics Inc. (Doty & Laing, 2003; Doty et al., 1986; Doty et al., 1995)). His threshold was -4.75 , exactly at the mean for his age group.

Neuropsychological testing

The neuropsychological results show that the patient has normal function in a comprehensive neuropsychological battery (see supplementary material). However, a lower score emerged in object decision (from the VOSP), total responses from d2, and interference (from Stroop color and word test) (Table S1).

TABLE 1
RG and control group (mean \pm SEM) rating of the different odors used in the Smell Identification Test (SIT)

Odor	Pleasantness rating		Intensity rating	
	RG	Control	RG	Control
Pizza	83*	48 (\pm 6.72)	69	62 (\pm 6.1)
Bubble gum	73	74 (\pm 3.2)	36*	73 (\pm 3.65)
Menthol	43*	77 (\pm 2.76)	16*	70 (\pm 12.62)
Cherry	44	60 (\pm 11.2)	53	43 (\pm 9.21)
Motor oil	27	40 (\pm 9.49)	41	39 (\pm 7.52)
Mint	67*	78 (\pm 2.24)	65	61 (\pm 9.67)
Banana	24*	76 (\pm 4.28)	56	60 (\pm 13.25)
Clove	76*	69 (\pm 3.1)	37*	65 (\pm 7.54)
Leather	21*	41 (\pm 7.47)	65	65 (\pm 11.67)
Coconut	84*	75 (\pm 3.8)	84*	59 (\pm 6.92)
Onion	16*	27 (\pm 2.49)	87*	69 (\pm 5.77)
Fruit juice	76	71 (\pm 4.76)	54	49 (\pm 11.27)
Talcum powder	30*	60 (\pm 7.63)	98*	66 (\pm 7.85)
Cheese	38	37 (\pm 6.84)	18	34 (\pm 11.31)
Cinnamon	76	71 (\pm 6.74)	68	59 (\pm 11.67)
Gasoline	15*	32 (\pm 7.86)	50*	67 (\pm 7.43)
Strawberry	48*	75 (\pm 6.22)	32	49 (\pm 15.69)
Cedar	48	51 (\pm 11.12)	65	43 (\pm 12.13)
Chocolate	57	52 (\pm 9.97)	60*	42 (\pm 8.9)
Apple	46	58 (\pm 11.29)	55*	30 (\pm 12.3)
Lilac	55*	81 (\pm 2.45)	35*	77 (\pm 6.72)
Turpentine	33*	62 (\pm 6.06)	56	42 (\pm 10.93)
Peach	60	64 (\pm 6.28)	37	40 (\pm 7.08)
Tire	8*	37 (\pm 13.07)	88*	66 (\pm 9.34)
Gherkin	64	60 (\pm 4.66)	62*	36 (\pm 12.15)
Pineapple	74	78 (\pm 4.82)	78	62 (\pm 14.73)
Raspberry	60	63 (\pm 9.63)	74	62 (\pm 8.66)
Orange	80	79 (\pm 5.49)	83*	59 (\pm 9.45)
Nut	38	48 (\pm 6.95)	79*	39 (\pm 5.89)
Beer	47*	66 (\pm 5.48)	35	27 (\pm 7.63)
Turpentine	46	43 (\pm 12.65)	90*	48 (\pm 12.1)
Grass	53	58 (\pm 7.11)	64	57 (\pm 11.25)
Smoke	22*	34 (\pm 3.01)	77*	68 (\pm 3.75)
Pine	36*	77 (\pm 2.9)	45	60 (\pm 7.73)
Artificial grapes	24*	55 (\pm 6.24)	29	40 (\pm 8.77)
Lemon	48	63 (\pm 10.60)	37	57 (\pm 10.61)
Soap	34*	63 (\pm 11.79)	58	60 (\pm 4.95)
Natural gas	12	22 (\pm 5.82)	69*	57 (\pm 5.66)
Rose	64*	74 (\pm 4.64)	74*	47 (\pm 11.23)
Peanut	42*	64 (\pm 7.91)	85*	69 (\pm 7.59)

*Indicates a difference of more than 2 SEM between RG and controls.

Face expression of emotion test

RG showed a specific disgust face expression recognition deficit in the 60 Faces and Emotion Hexagon Test part of the Facial Expression of Emotion: Stimuli and Tests (FEEST) (Young et al., 2002) (Table 2). The recognition scores for the other face emotion expressions (anger, fear, happiness, sadness, and surprise) were in the normal range (Table 2). Further, to control for whether being a

pathologist might be relevant to the reduced disgust scores of RG, we measured the disgust face recognition scores of four of RG's pathology colleagues. They were for the 60 faces test 7.25 ± 0.25 (mean \pm SEM) (compared to RG's score of 4) and for the hexagon test 18.25 ± 1.18 (mean \pm SEM) (compared to RG's score of 3). The evidence thus is that RG performed well below the norm and well below the performance of his colleagues on the identification of the face expression of disgust. Thus, in the patient RG, a unilateral right amygdala lesion was associated with a specific impairment in the identification of the face expression of disgust. The performance was normal at all the other face expressions in the test.

DISCUSSION

The main findings in this patient were that the gourmand status was associated with a lesion to the right amygdala and temporal pole; that the taste thresholds were normal; that RG had an unusual liking for salt (NaCl) at low and moderate concentrations, or, to put it another way, did not dislike the taste of salt as much as age-matched controls; that this also occurred for MSG; that there were no clear olfactory differences from what might be expected; and that there was a marked reduction in the ability to detect face expressions of disgust. In addition, it is noted that the tissue associated with the cavernous sinus and indicated in Figure 1d was treated with radiotherapy in 2010 and that the gourmand status declined after that. We consider these findings further.

This patient had a clearly localized lesion in the right amygdala (i.e., unilaterally), with damage also to the right temporal pole cortex just anterior to the amygdala (Figure 1). The hippocampus and insula were remarkably intact. The lesion in the right amygdala and temporal pole cortex is thought to be due to the large fibroblastic meningioma in the minor wing of the sphenoid bone that was removed in 1997. His gourmand status, which appeared four years before surgery, can be attributed to the slow growing rate typical of this type of fibrous meningioma since the previous history does not support an explanation based in having being exposed to refined gastronomic habits. Thus, it is feasible that the fibrous meningioma was pressing on the right temporal lobe several years before being diagnosed following the olfactory hallucinations. This finding has great relevance since publications on the

TABLE 2
Ekman 60 faces and Emotion Hexagon results

<i>Emotional test</i>	<i>Emotion</i>	<i>Score</i>	<i>Manual cut-off age 41–60</i>	<i>Manual mean age 41–60</i>
Ekman 60 Faces Test (max = 10)	Anger	5	5	8.17
	Disgust	4*	6	8.77
	Fear	8	4	7.23
	Happiness	10	9	9.84
	Sadness	9	6	8.53
	Surprise	9	6	8.61
	Grand total (max = 60)	45	43	51.20
Emotion Hexagon Test (max = 20)	Anger	19	13	18.10
	Disgust	3*	13	17.63
	Fear	17	10	18.71
	Happiness	20	18	16.15
	Sadness	20	13	19.63
	Surprise	16	14	18.31
	Grand total (max = 120)	95	92	108.10

* Indicates where the scores of the patient are below the cut-off for age-matched controls. Max indicates the maximum score attainable on a test.

“Manual cut-off” and “manual mean” are the cut-off and the mean provided by the FEEST manual, respectively.

gourmand syndrome have not described damage in the amygdala in particular, though right temporal lobe damage is frequently reported, and in some of the previous cases there has been damage more posteriorly in the temporal lobe, e.g., in the posterior ventral insula (Cockrell, 1998; Kurian et al., 2008; Myslobodsky, 2003; Regard & Landis, 1997). Consistent with the lack of damage in RG to the anterior insula where the primary taste cortex is located (Pritchard, Hamilton, Morse, & Norgren, 1986; Rolls, 2005, 2008, 2011; Rolls & Grabenhorst, 2008), taste thresholds were normal. It is possible that in this patient some pressure on the medial temporal lobe from the small growth associated with the cavernous sinus (see Figure 1d) contributed to the gourmand status, for after radiotherapy that reduced the size of this growth, the gourmand status declined. Of course, other factors associated or not with the radiotherapy may also have made a contribution to the change of status.

The reduction in the dislike for salt solutions at most concentrations (except for the highest) (Figures 3 and 4) was a marked difference in RG from controls. This is an affective difference from controls in taste responsiveness that is associated with damage to the right amygdala. We do not know what role, if any, this played in his gourmand status, and it will be very interesting to follow up in other patients to investigate whether this is common. What we did not find was any particular hyper-hedonia for any aspect of taste or odor, and this reduction in the dislike for most

salt solutions was the closest finding in this direction. However, we do note that at the time of testing (November 2011) RG's gourmand interest in food had declined, perhaps associated with the further treatment he received in 2010, which may have alleviated some previous effects on the temporal lobe due to the growth in the cavernous sinus. For that reason, it would be informative to know the results of taste and olfactory tests along the lines of those described here performed in other patients with gourmand status. It is known that amygdala neurons respond to a wide range of tastes (Kadohisa, Rolls, & Verhagen, 2005a) and are especially concerned with oral texture (Kadohisa et al., 2005a; Kadohisa, Rolls, & Verhagen, 2005b), and that the human amygdala is as well activated by a pleasant sweet taste as by an unpleasant salt taste (O'Doherty, Rolls, Francis, Bowtell, & McGlone, 2001); so a deficit in taste processing selective for salt, if it is related to amygdala damage, is somewhat surprising.

Another finding that seems consistent in some way with the reduction in the dislike found for a taste was the difficulty in recognizing the face expression of disgust (Table 2). RG reports not having realized it until he underwent the neuropsychological assessment. If the strong selective impairment in recognizing the face expression of disgust is a result of the damage to the right amygdala, that is of great interest, for, though amygdala lesions are known to cause face expression recognition deficits, the deficit is usually said

to be selective for fear face expressions (Adolphs et al., 2005; Calder et al., 1996; Phelps & LeDoux, 2005), whereas the insula is activated by disgust face expressions (Phillips et al., 2004) and damage to the insula (and basal ganglia) may impair face expression identification (Calder, Keane, Manes, Antoun, & Young, 2000; Kipps, Duggins, McCusker, & Calder, 2007). The present finding suggests that the amygdala with its face processing neurons (Leonard, Rolls, Wilson, & Baylis, 1985) may not be concerned only with fear. However, the present person had a right unilateral amygdala lesion and also had unilateral damage to the right temporal pole cortex, and so was different in these respects from previous patients that have been investigated. In relation to the impairment in identifying face expressions of disgust, it will be of interest to investigate whether other people with the gourmand syndrome associated with right temporal lobe damage also show this change in face emotion expression processing.

In some fMRI studies, amygdala activation has been produced by all emotions (Derntl et al., 2009; Tettamanti et al., 2012), and disgust has been associated with the orbitofrontal and insula cortex (Jehna et al., 2011) and disgust-related conditioning with the insula (Klucken et al., 2012). However, it must be remembered that below the primary taste cortex in the anterior insula is a region that is probably autonomic/visceral cortex (Baylis, Rolls, & Baylis, 1995; Critchley, 2005). A visceral efferent system might be expected to be especially active if it receives inputs from elsewhere in the brain specifying that a stimulus evokes disgust and an autonomic response (Grabenhorst & Rolls, 2011; Rolls, 2008; Rolls & Grabenhorst, 2008). If the evoking stimulus was a taste stimulus, the inputs might be from the taste insula, orbitofrontal cortex, or amygdala. If the evoking stimulus was visual, the inputs might come via the orbitofrontal cortex and anterior cingulate cortex to the visceral insula. If the input was somatosensory (e.g., an unpleasant oral texture), the input might come from the taste cortex (which has oral texture representations) and from the orbitofrontal cortex, which also has oral texture representations (Grabenhorst & Rolls, 2011; Rolls, 2008; Rolls & Grabenhorst, 2008). However, in these cases, the insula might be mainly related to disgusting stimuli because of its relation to autonomic output response rather than to a specific role in decoding visual stimuli that portray the expression on a face. The visual decoding

of face expression appears to be performed in the cortex in the superior temporal sulcus (Hasselmo, Rolls, & Baylis, 1989) and in the orbitofrontal cortex (Hornak, Rolls, & Wade, 1996; Rolls, Critchley, Browning, & Inoue, 2006), and neither of these areas is specific for a particular face expression but is involved in most face expressions (Hornak et al., 1996). With respect to the amygdala, abnormal right amygdala reactivity to facial expressions has been reported in eating disorders. Patients with bulimia nervosa exhibit a decreased neural response to angry facial expressions in the right amygdala (and a decreased response to both anger and disgust in the precuneus) (Ashworth et al., 2011). Patients with anorexia nervosa show increased activity in the right amygdala to photographs depicting food and nonfood items, and the signal correlates negatively with disgust ratings (Joos et al., 2011).

In conclusion, we have described the first systematic study we know of the taste and olfactory status of a patient with a gourmand syndrome associated with damage to the right temporal lobe. The main findings in this patient were that the gourmand status was associated with a lesion to the right amygdala and temporal pole, and with pressure on the right medial temporal lobe; that the taste thresholds were normal; that RG had an unusual liking for salt (NaCl) at low and moderate concentrations, or, to put it another way, did not dislike the taste of salt as much as age-matched controls; that there were no clear olfactory differences from what might be expected; and that there was a marked reduction in the ability to detect face expressions of disgust. We believe that it will be of interest to perform tests for olfactory and taste changes in other gourmand patients with damage to the medial temporal lobe and to investigate whether other gourmand patients also have impairments in face expression identification. These are of some interest in the present patient with right amygdala and temporal pole damage, for the change was in the identification of face expressions of disgust. This change in the identification of a face expression normally associated with an unpleasant taste may fit in some way with the gourmand status. The fact that it was disgust and not fear face expression identification that was impaired implies that amygdala damage is not related uniquely to an impairment in the identification of face expressions of fear and that insular cortex lesions are not related uniquely to an impairment in the identification of face expressions of disgust.

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Supplementary material

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